

## Ominous comorbidities: Small ventricular septal defect and warm autoimmune hemolytic anemia

Sir,

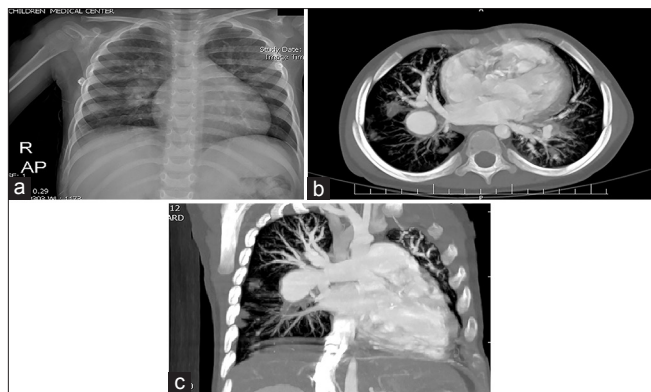
A 4-year-old asymptomatic girl was brought to the outpatient clinic for a checkup visit. She was clinically followed for a known diagnosis of small ventricular septal defect (VSD) by a local pediatric cardiologist since early infancy. She had a history of warm autoimmune hemolytic anemia (WAHA) for 1 year ago which was successfully controlled by prednisolone and cyclosporine at a dose of 2 mg/kg/day and 2 mg/kg/day, respectively. She had a mildly cushingoid face and was in good general condition with normal growth and development.

However, findings on her cardiac examination were not explained with her history of the small VSD. P2 component of the second heart sound was loud, and a regurgitant murmur of 3/6 was heard at the lower left sternal border. Complete two-dimensional echocardiography revealed a very large, about 4 cm × 4 cm vegetation extending from the tricuspid valve to the pulmonary valve, producing moderate

tricuspid regurgitation with a pressure gradient of 90 mmHg and mild pulmonary stenosis and regurgitation. There was a small VSD which was almost sealed by the large vegetation [Video Clips 1 - 3].

Electrocardiogram showed right ventricular hypertrophy, and chest X-ray revealed a round consolidation in the right lung [Figure 1a].

Computed tomographic angiography (CT) of the chest showed multiple thromboembolism in the right and left lung with aneurysmal dilation of the right pulmonary artery [Figure 1b and c and Video Clips 4 and 5]. The child was admitted. Intravenous (IV) antibiotics were started with the diagnosis of bacterial endocarditis. Blood cultures obtained before antibiotic therapy were positive for group B streptococcus viridians with sensitivity to penicillin and ampicillin. Since there was no change in the size of the vegetation after 6 weeks of IV antibiotic therapy, the patient underwent elective cardiac surgery. The huge vegetation with the calcification and abscess formation inside it was removed. Tricuspid and



**Figure 1:** (a) Chest X-ray of the child at the time of admission shows a round consolidation in the right lung. (b and c) are the computed tomographic angiographic images which indicate the dilation and cutoff the pulmonary blood flow in the branch of right pulmonary artery due to thromboembolism. The short-axis cut of the right pulmonary artery has produced a circular appearance resembling the “moon in the sky”

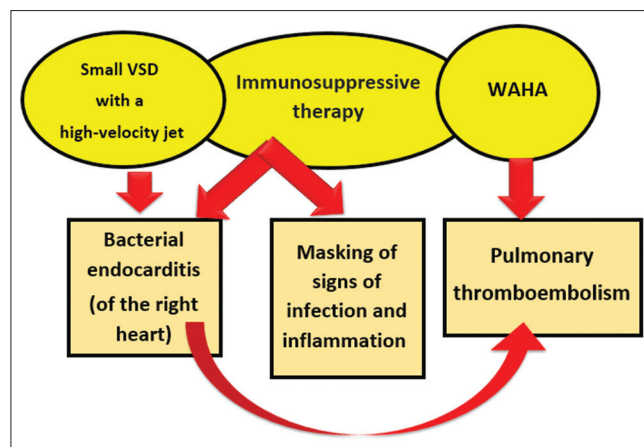
pulmonary valves were repaired and thrombectomy of the pulmonary artery branches were done. The small 3-mm VSD was closed by simple sutures. Postoperative IV antibiotics were continued for 4 weeks. Despite advice of the pediatric cardiologist on the necessity of continued IV antibiotic therapy, the parents took the child to home. Two weeks later, the child was brought to the emergency department in a seriously ill condition with respiratory distress. Her echocardiographic examination revealed recurrence of the vegetation in the right ventricle. Cardiopulmonary CT angiography revealed massive pulmonary thromboembolism. The child died of fatal pulmonary hemorrhage.

Closure of small VSD is not recommended in the literature.<sup>[1]</sup> However, certain forms of idiopathic WAHA predisposes the child to thromboembolic events including venous thromboembolism and pulmonary thromboembolism.<sup>[2-5]</sup> In this case, the triad of “small VSD,” “WAHA,” and “immunosuppressive treatment” predisposed the child to clinically masked bacterial endocarditis and pulmonary embolism. Prednisolone and cyclosporine disguised the clinical manifestations of infection and inflammation in this child [Figure 2].

WAHA and VSD are ominous comorbidities. Closure of even small-sized VSDs is strongly recommended in a child with WAHA. To close or not to close a VSD, it is the “whole” of the patient that counts not simply the size of the “hole” in his/her heart.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for



**Figure 2:** This scheme shows the interactions between the ominous comorbidities of small ventricular septal defect and warm autoimmune hemolytic anemia, a hemolytic disease that predisposes to venous and pulmonary thromboembolic events

his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

**Elaheh Malakan Rad**

Department of Pediatrics, Children's Medical Center  
(Pediatric Center of Excellence), Tehran, Iran  
E-mail: erad@tums.ac.ir

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